



Commonly Encountered Congenital Heart Disease in the Adult

Sabrina Phillips, MD FACC FASE
Mayo Clinic Congenital Heart Disease Center

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Atrial Septal Defect

- Second most common congenital defect recognized in adulthood
- Symptoms progressive
- Physical exam findings subtle



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Atrial Septal Defects

- Secundum
- Primum
- Sinus Venosus
- Unroofed Coronary Sinus



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Atrial Septal Defect

- Echo Diagnosis and Evaluation
 - Location of Defect
 - Right sided chamber size and function
 - Estimation of PA pressure
 - Tricuspid Regurgitation
 - Other Lesions
 - Repair Options



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Secundum Atrial Septal Defect

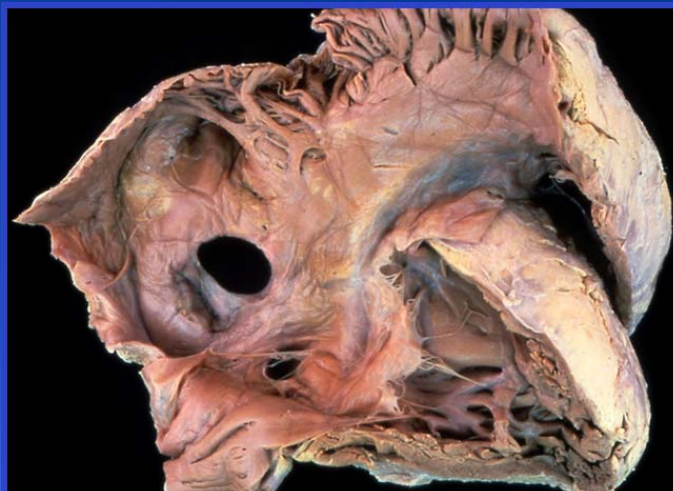
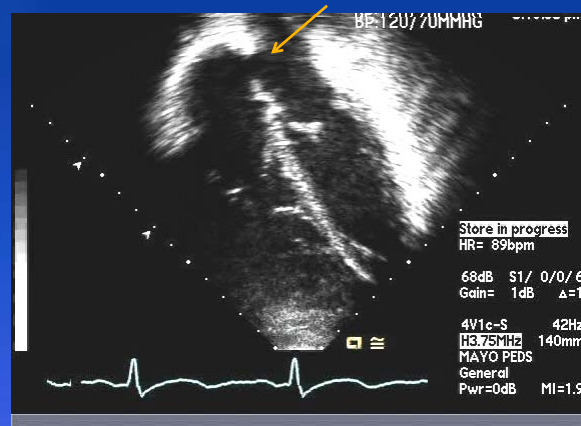


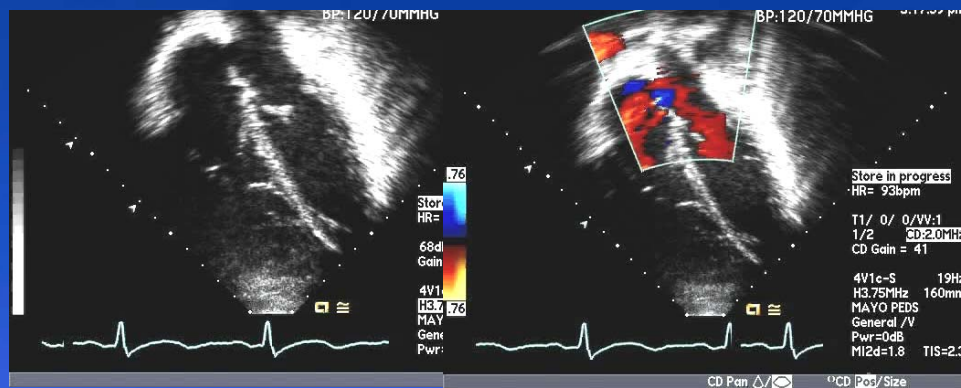
Image Courtesy of Dr. Bill Edwards

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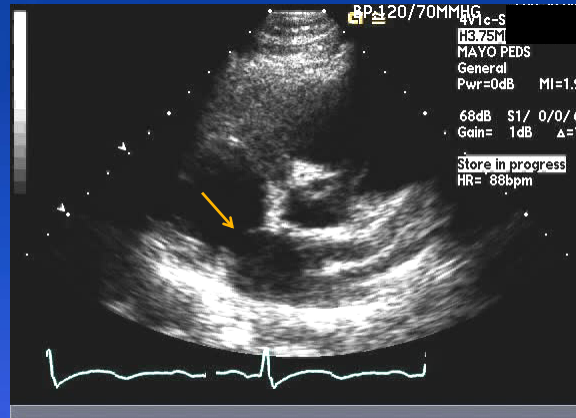
Apical 4 Chamber Imaging



Apical 4 Chamber Imaging

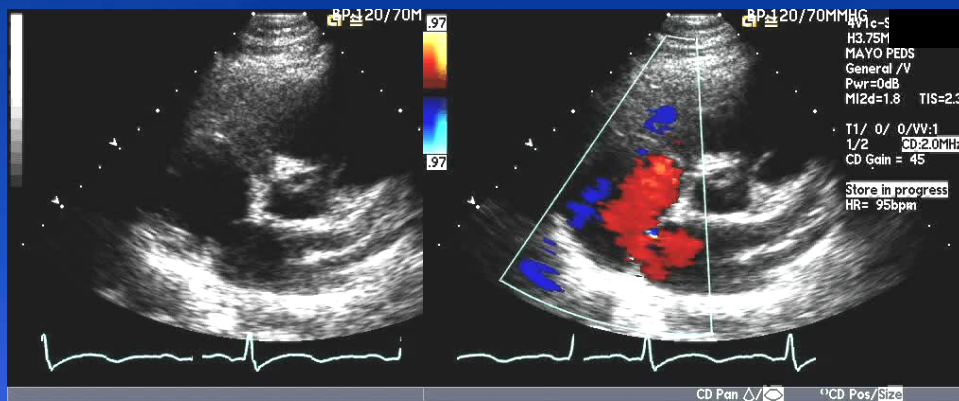


Parasternal Short Axis Imaging



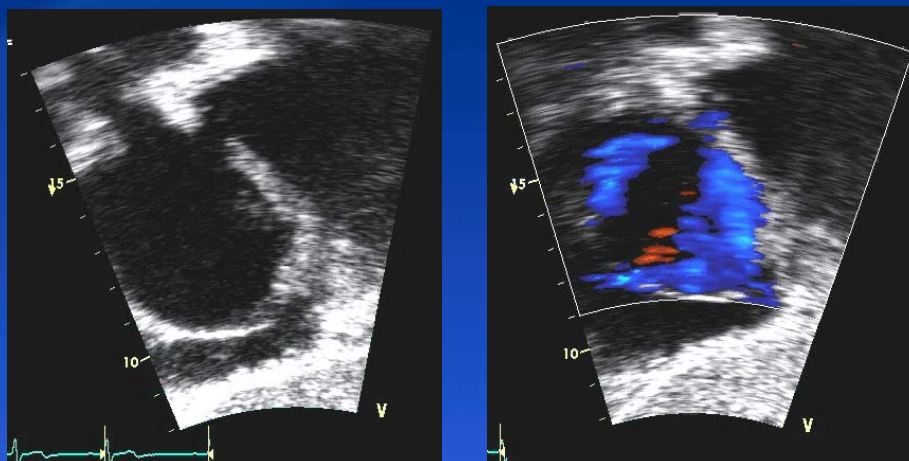
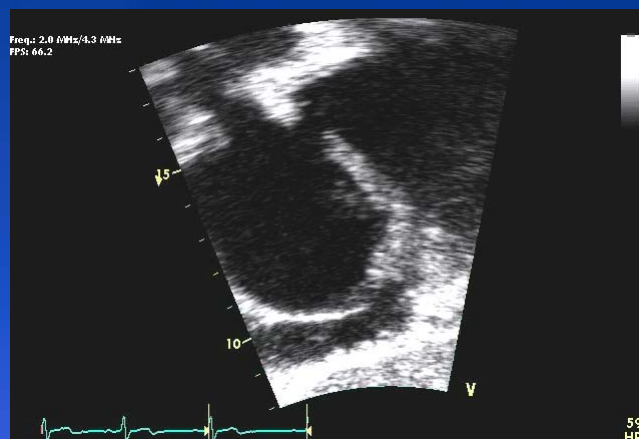
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Parasternal Short Axis Imaging



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Subcostal Imaging



Primum Atrial Septal Defect

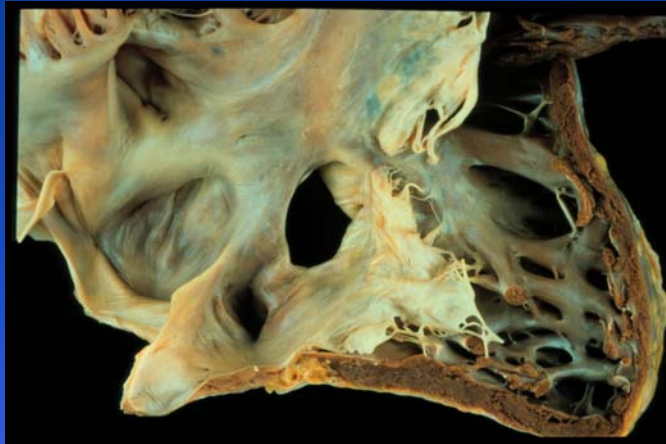
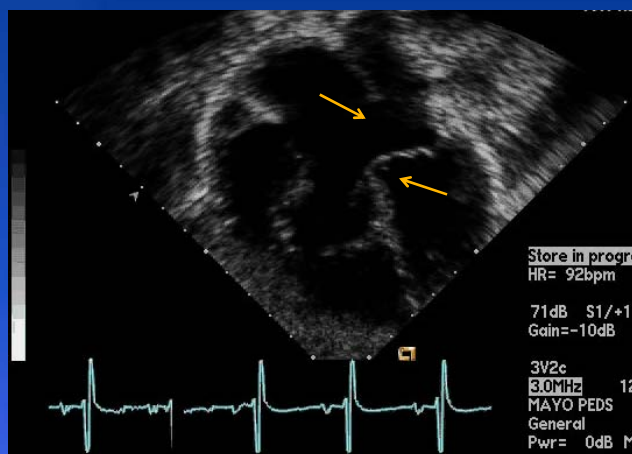


Image Courtesy of Dr. Bill Edwards

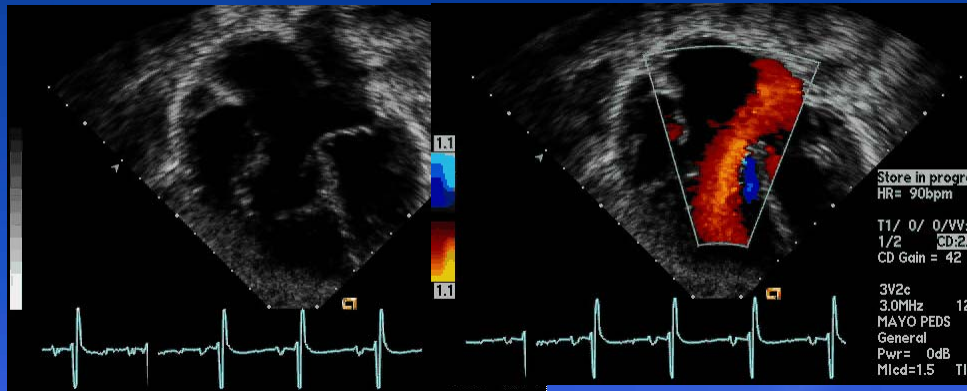
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Apical 4 Chamber Imaging



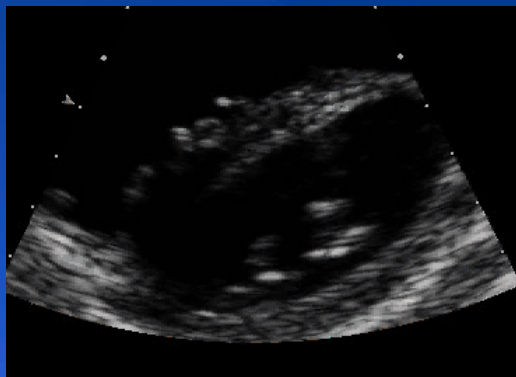
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Apical 4 Chamber Imaging

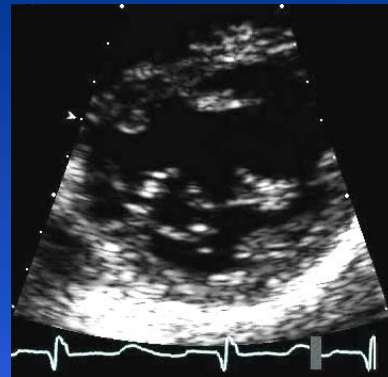


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Valvular Abnormalities Associated with Primum ASD



Cleft Mitral Valve

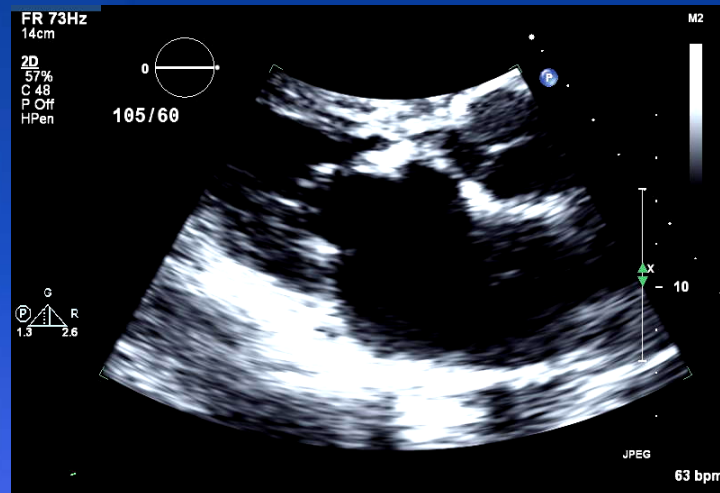


Double Orifice
Mitral Valve



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LVOT Elongation, Narrowing, Anomalous Chords

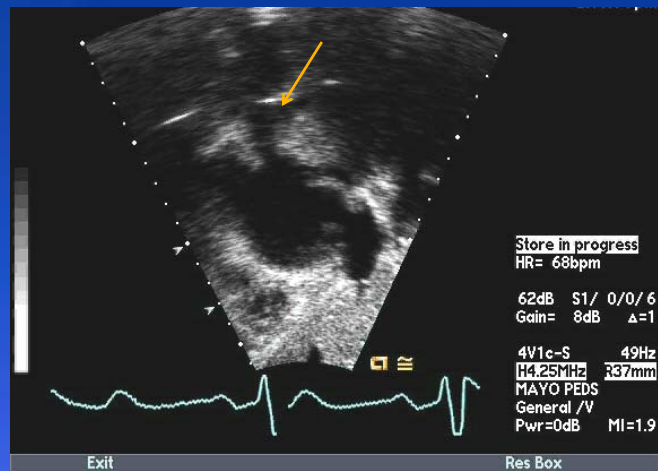


Sinus Venosus Atrial Septal Defect



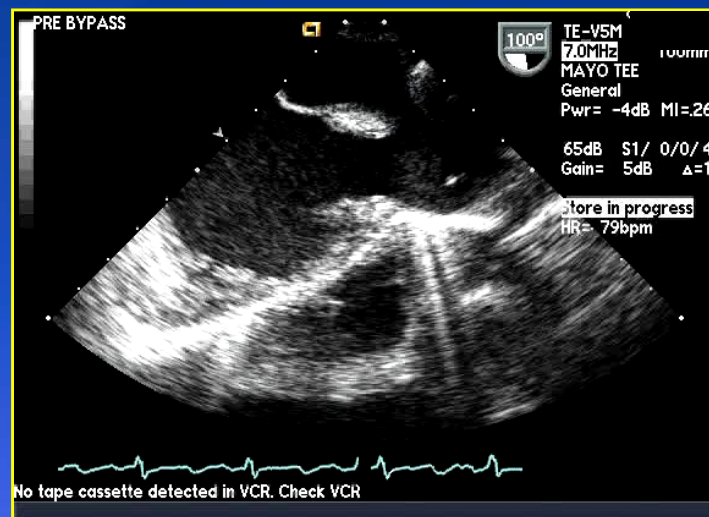
Image Courtesy of Dr. Bill Edwards

Subcostal Imaging



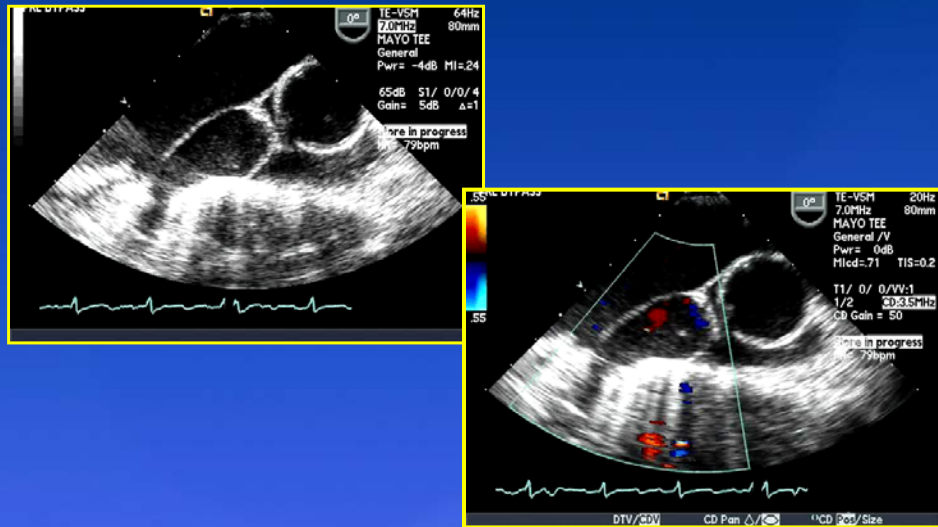
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TEE Imaging

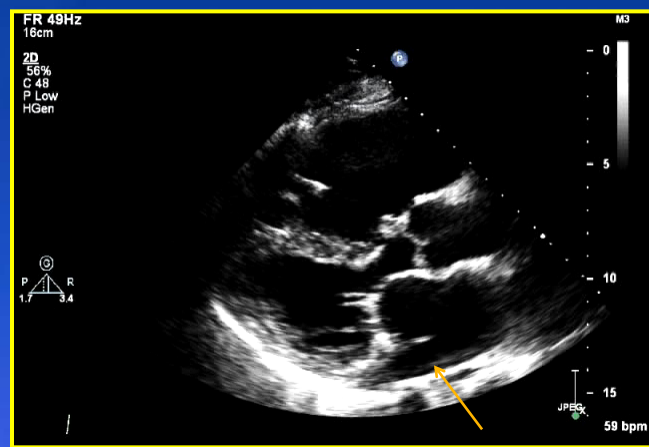


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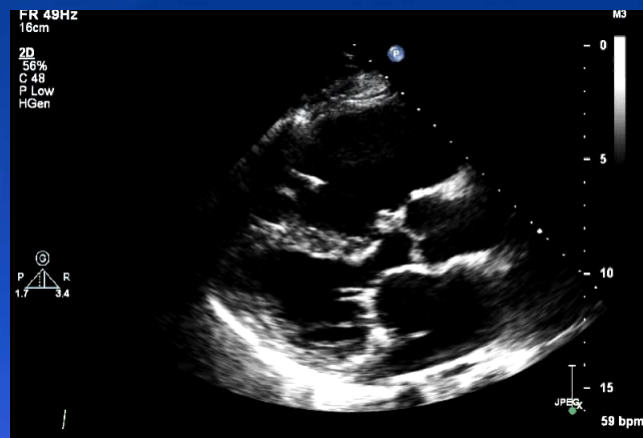
Anomalous Right Pulmonary Vein



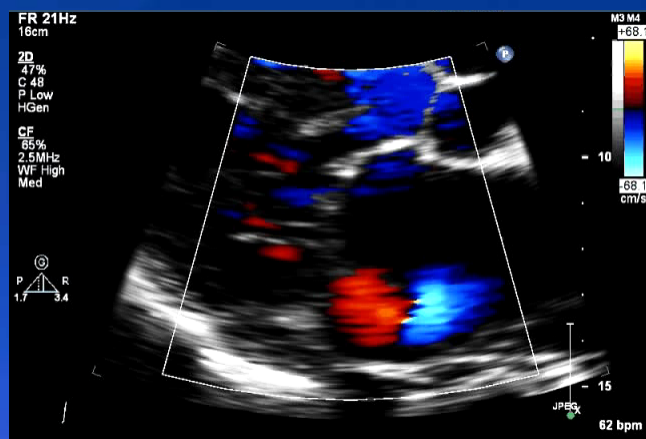
Coronary Sinus Atrial Septal Defect



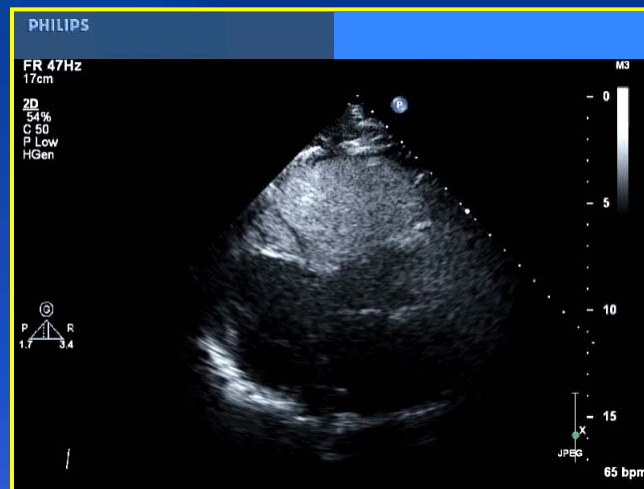
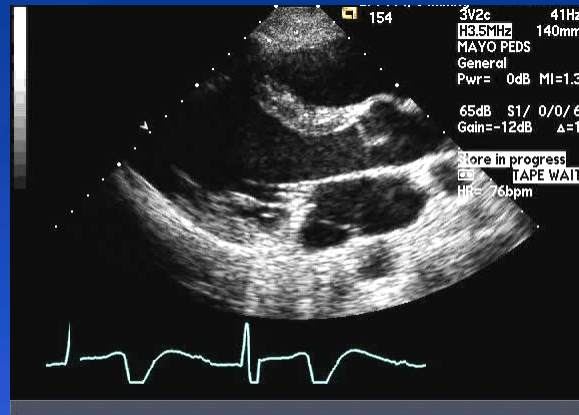
Coronary Sinus Atrial Septal Defect



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Partial Anomalous Pulmonary Venous Return/Connection



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Variants of Partial Anomalous Pulmonary Venous Connection

- Right pulmonary venous anomalies are most common
- Left pulmonary venous anomalies only comprise 4% of PAPVC
- Scimitar syndrome 3% of PAPVC
- Connections to the CS exceedingly rare
- Bilateral PAPVC occurs, but rare



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PAPVC Physiology

- Left to right shunt
- Right chamber volume overload and dilatation
- Single anomalous veins – low risk of hemodynamic compromise
- Less than 50% shunt – rare to have symptoms in childhood



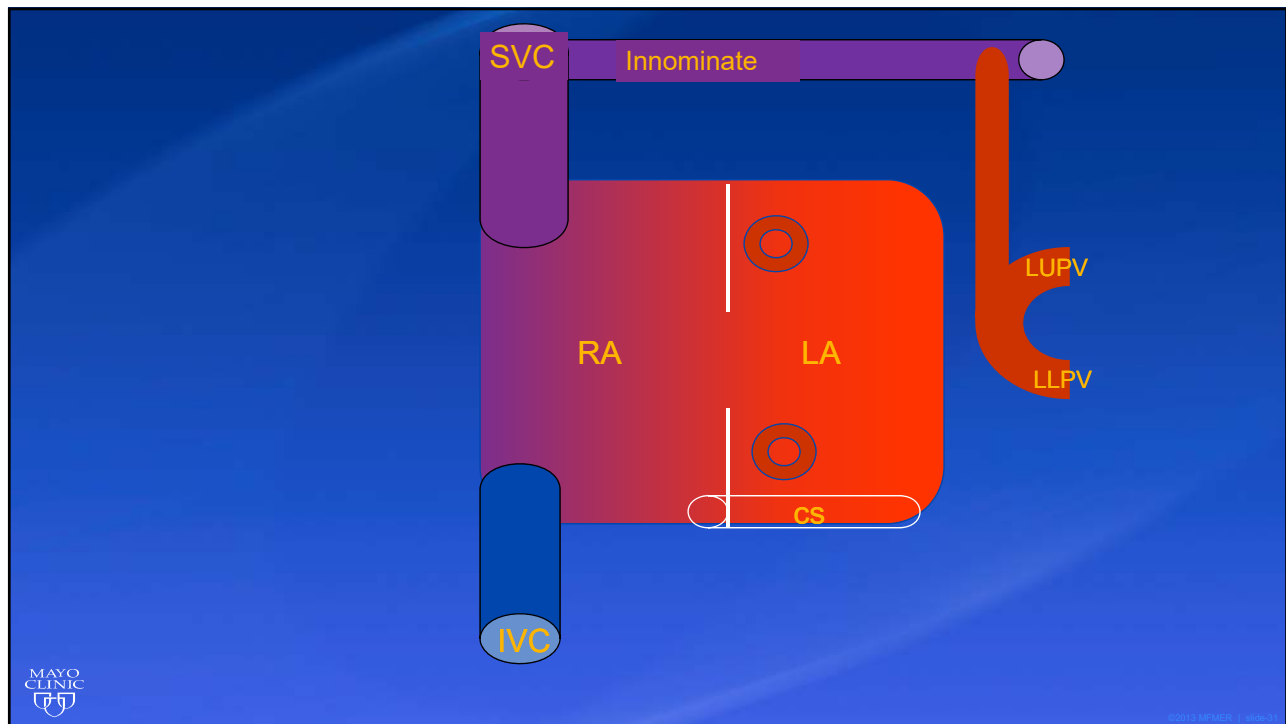
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ECHO Evaluation of PAPVC

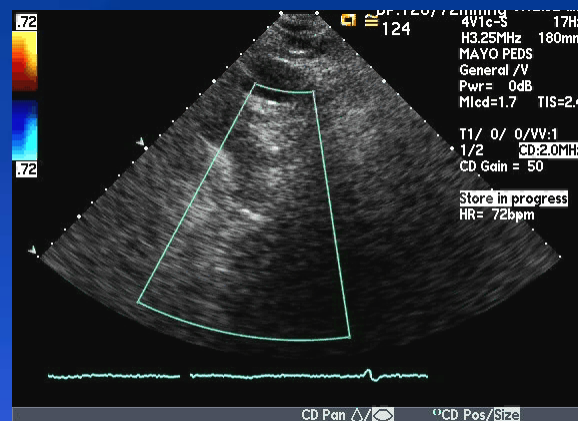
- Type of connection
- Associated anomalies
- Right chamber size
- Right ventricular function
- Pulmonary artery pressure

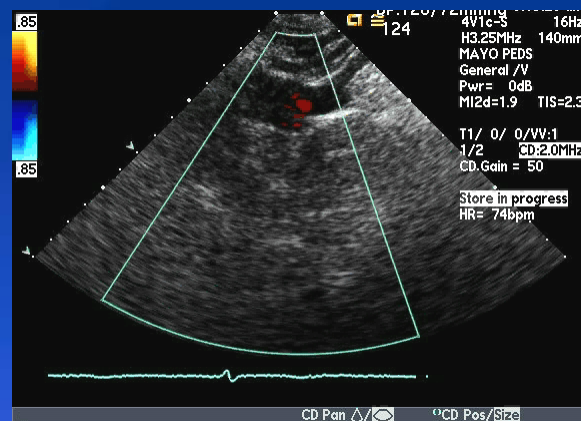
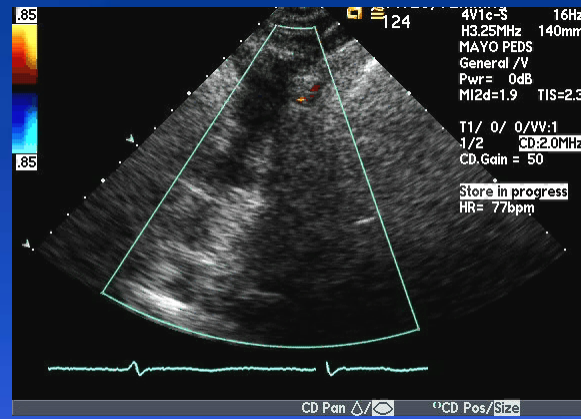


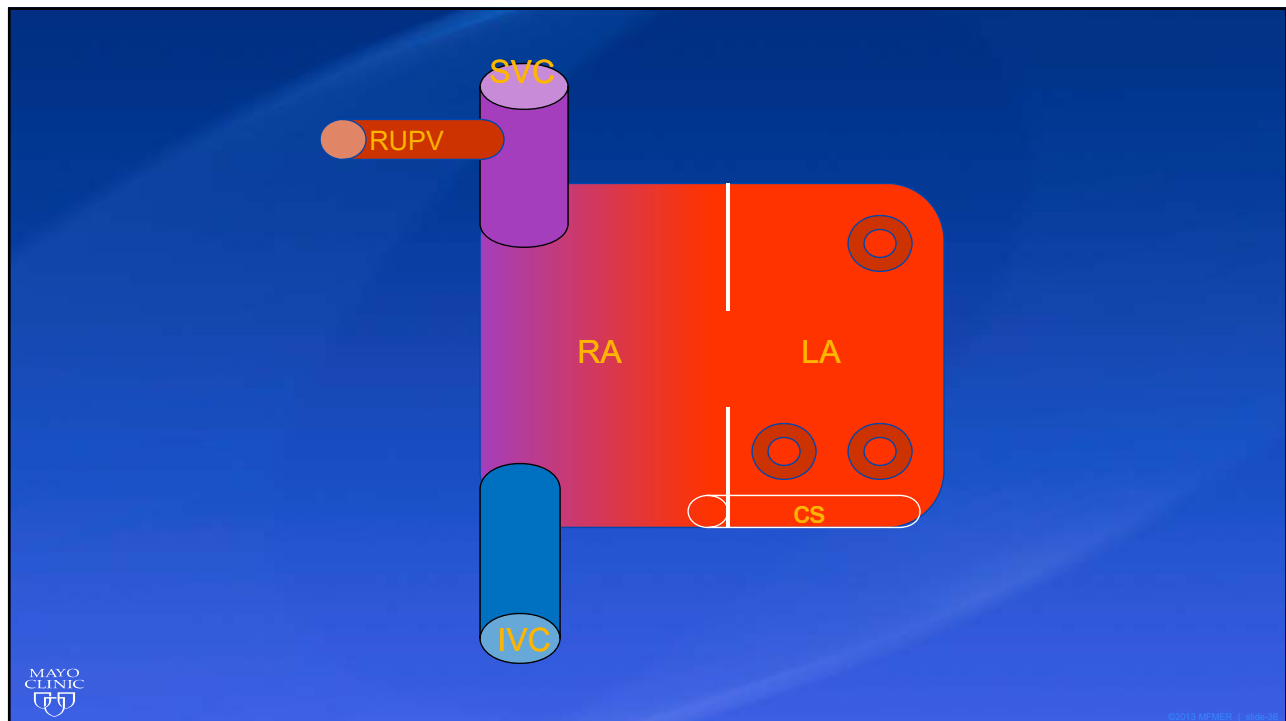
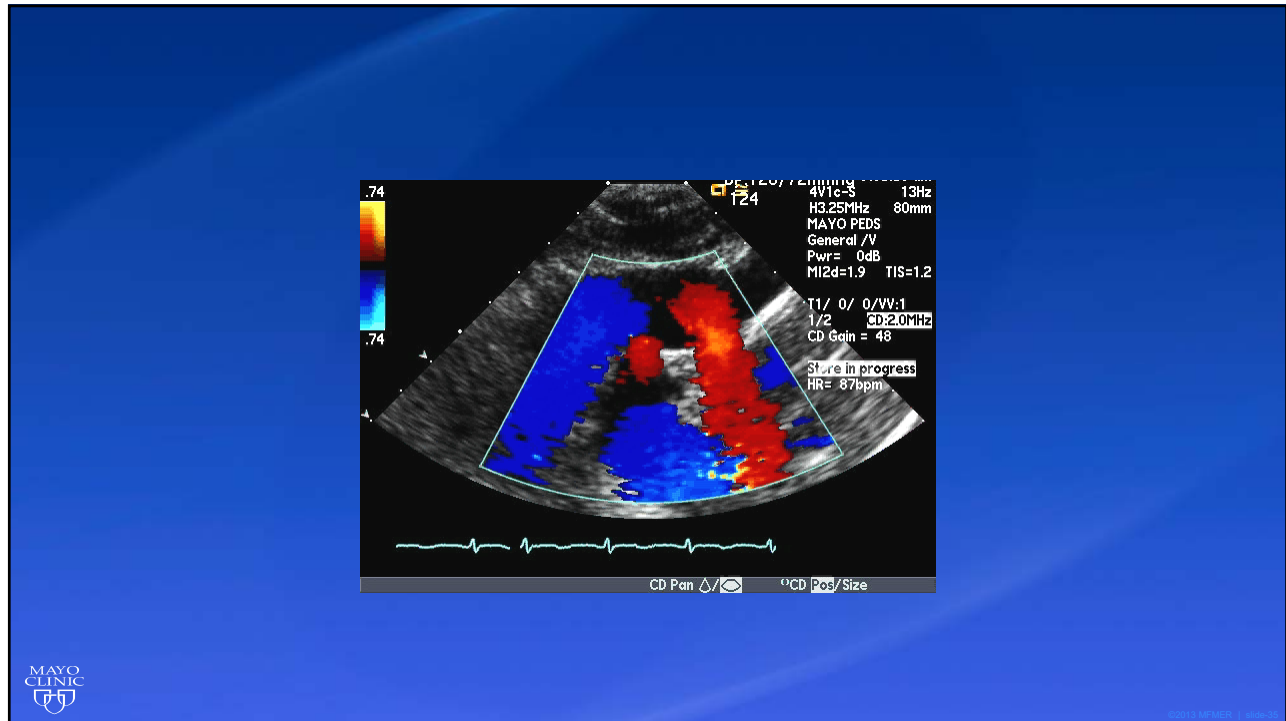
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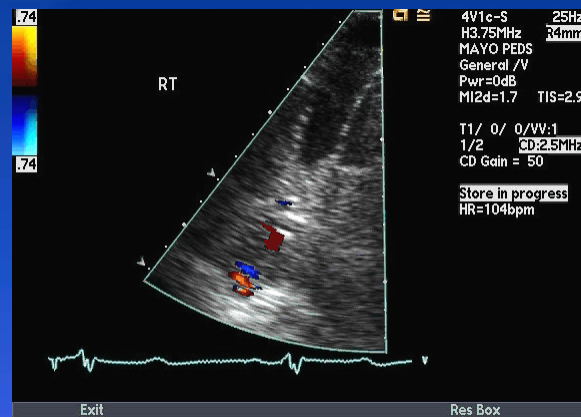
Vertical Vein



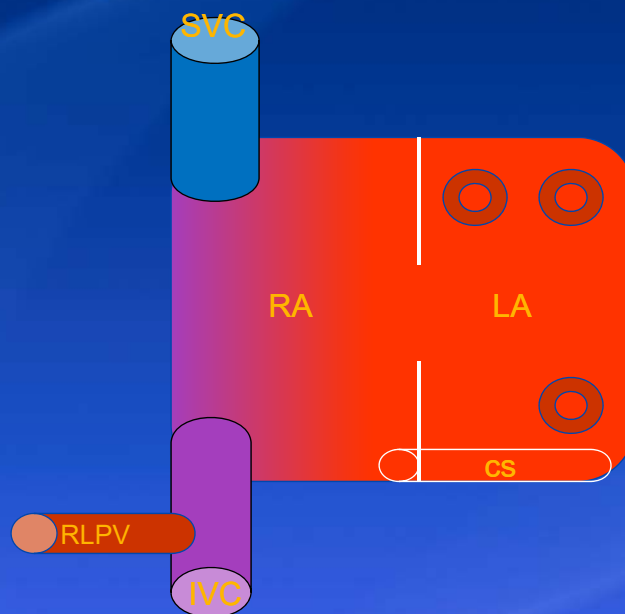




Right Pulmonary Vein to SVC

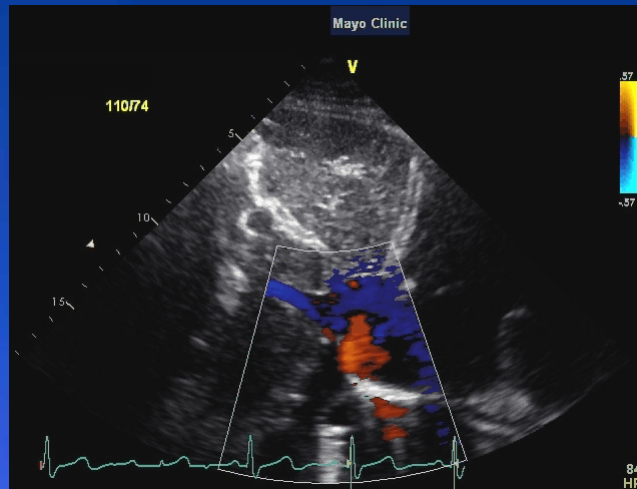


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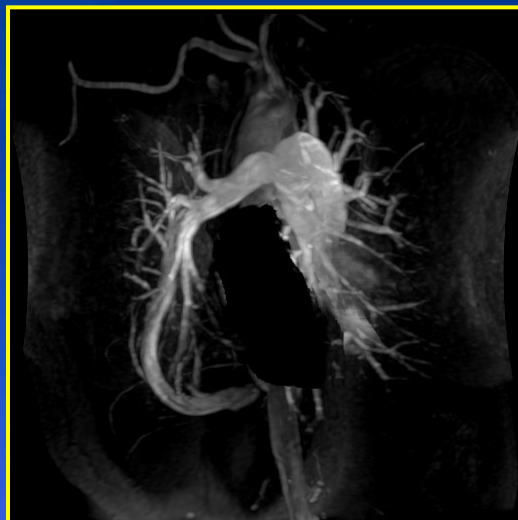


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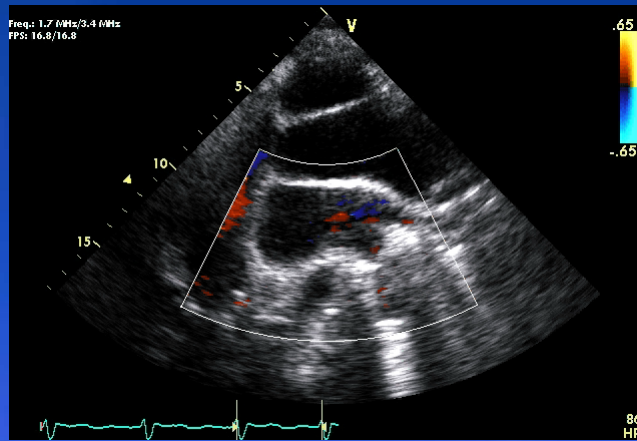
Scimitar Syndrome



MRA



Suprasternal Notch Coronal View ("Crab")



Ventricular Septal Defects

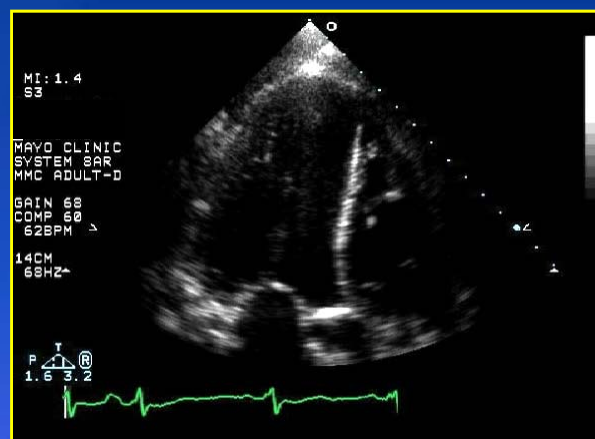
Echo Evaluation of VSDs

- Location
- Size
- Involvement of other structures
- *Left* ventricular and *left* atrial size
- Estimated right ventricular systolic pressure
- Associated anomalies

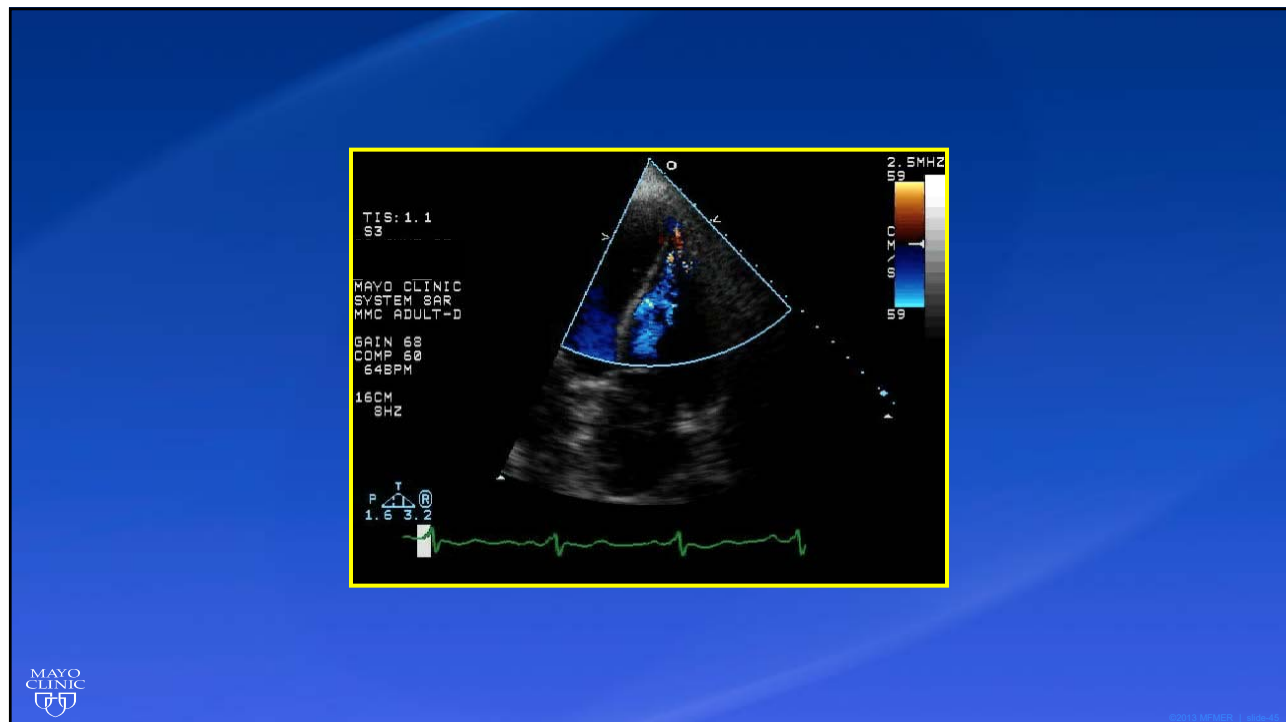


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Post- MI VSD



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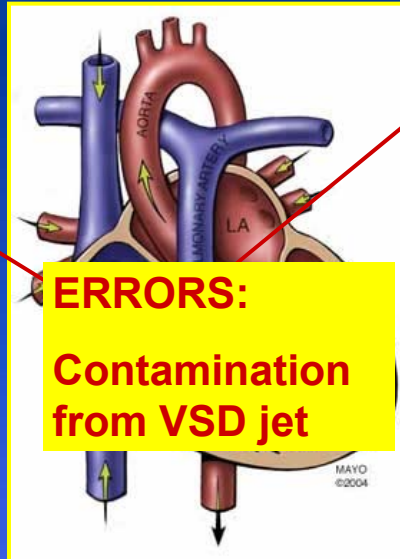


VSD Caveats

- The VSD jet may contaminate the TR signal
- Patients with high RV pressures may not have much color flow

Pulmonary Hypertension?

TR estimates
RV pressure

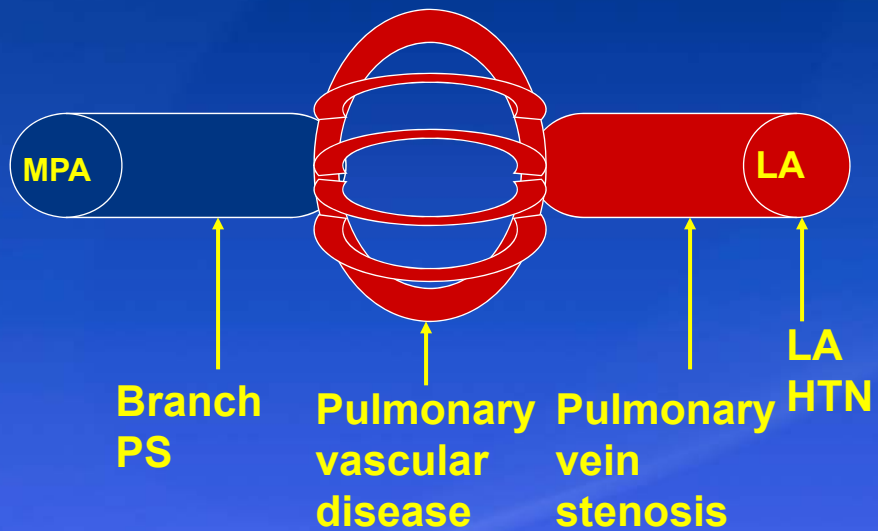


Outflow
Obstructions:

1. PS
2. Double
chamber
RV



Pulmonary Hypertension



$$\text{PAP} = \text{QP} \times \text{PVR}$$

PAP: Pulmonary artery pressure

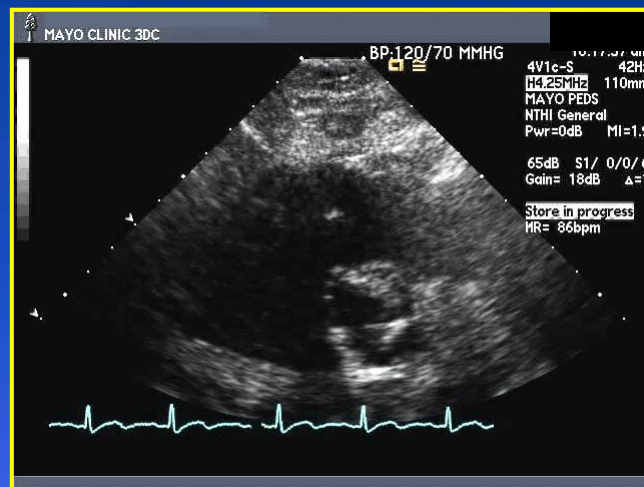
QP: Pulmonary blood flow

PVR: Pulmonary vascular resistance

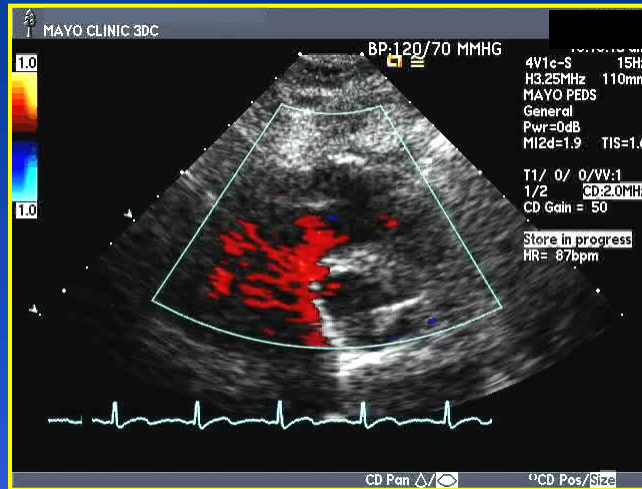


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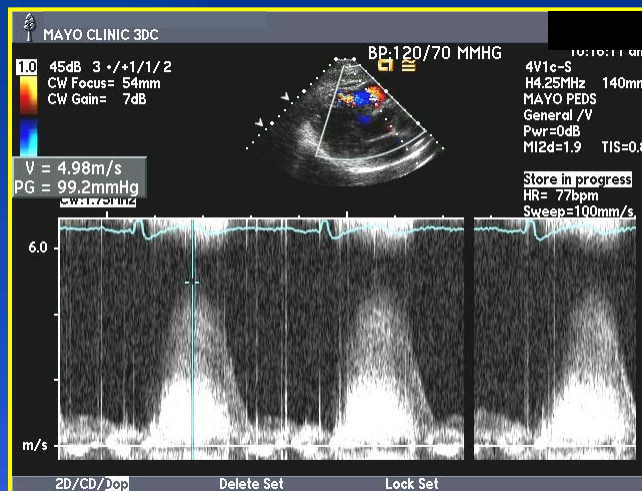
Double Chambered RV



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Indications for Closure

- Large VSD (left heart enlargement, QP/QS > 1.5) without irreversible pulmonary vascular disease
- Aortic valve prolapse with progressive regurgitation
- RV outflow tract obstruction
- Recurrent endocarditis



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Atrial Septal Defect vs. Ventricular Septal Defect

ASD

- Increased pulmonary blood flow
- Primarily volume load
- Low incidence of pulmonary hypertension in adulthood

VSD

- Increased pulmonary blood flow
- Primarily pressure load
- High incidence of pulmonary hypertension in adulthood



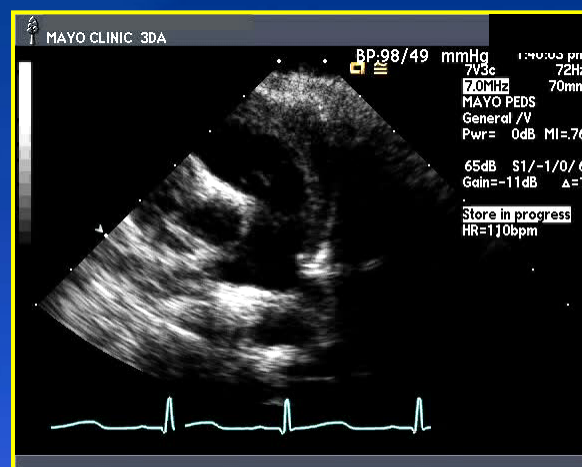
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Patent Ductus Arteriosus

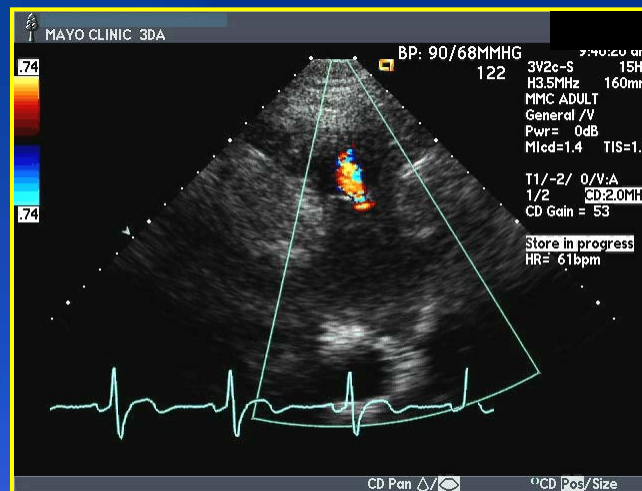
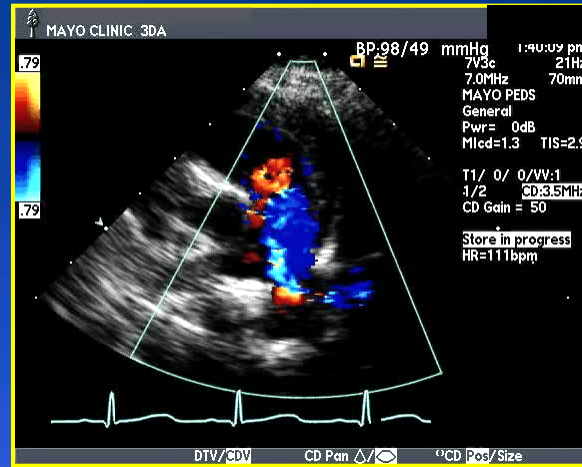
- Left Heart Enlargement
- Pulmonary hypertension common if the PDA is large – may not see a shunt on echo (equal pressures)



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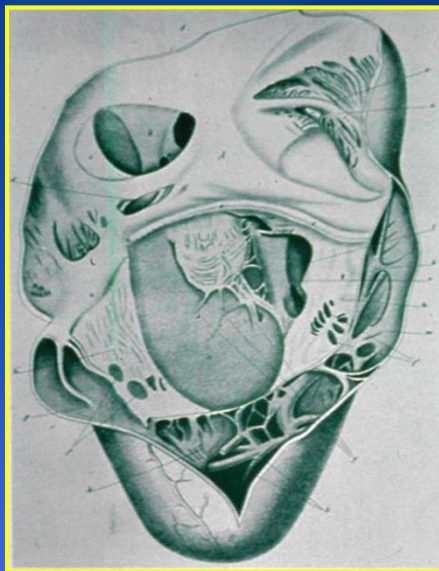
QP/QS

~~Flow Diameter $\times 0.785 \times \text{RVO1}$
Flow Diameter² $\times 0.785 \times \text{LVOT}$~~



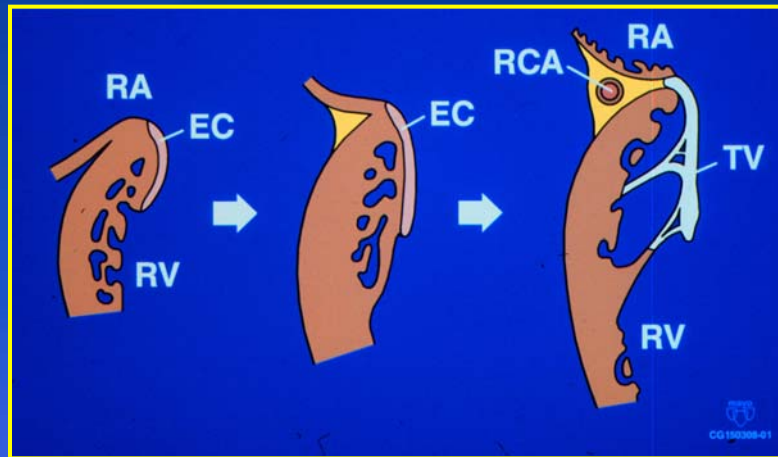
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Ebstein Anomaly



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Normal Delamination of the TV from the RV Myocardium

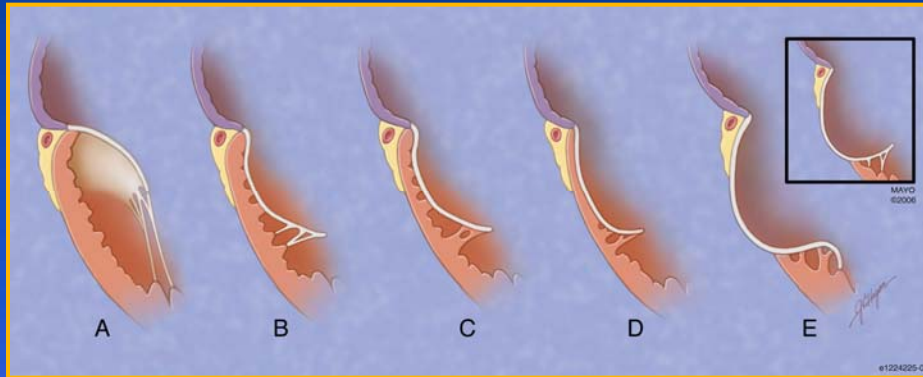


Failed Delamination results in ...



- adherence of leaflets to underlying RV myocardium
- displacement of the anular hinge points

Failure of Delamination From the Myocardium

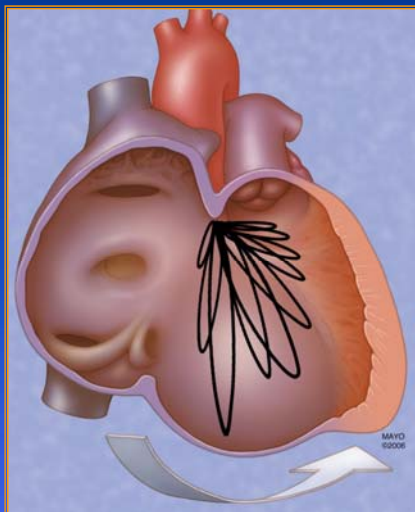


Spectrum with Infinite Variability



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Displacement Apically AND Toward the Right Ventricular Outflow Tract



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Echocardiographic Diagnosis

- Apical displacement of the septal leaflet of the tricuspid valve > 8mm/m2
- Right sided chamber enlargement with “atrialized” RV
- Tricuspid valve regurgitation – often appears laminar
- Elongated, tethered anterior TV leaflet



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Apical Displacement: 20 mm



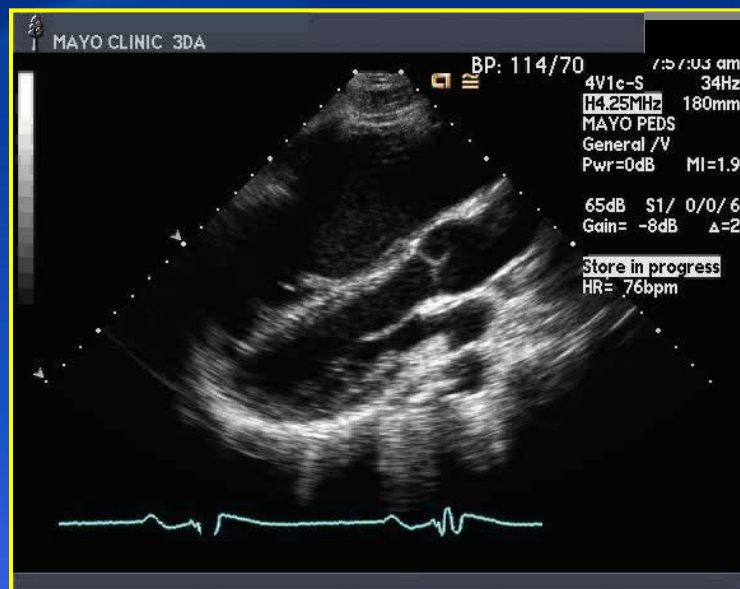
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Ebstein Anomaly Associated Lesions

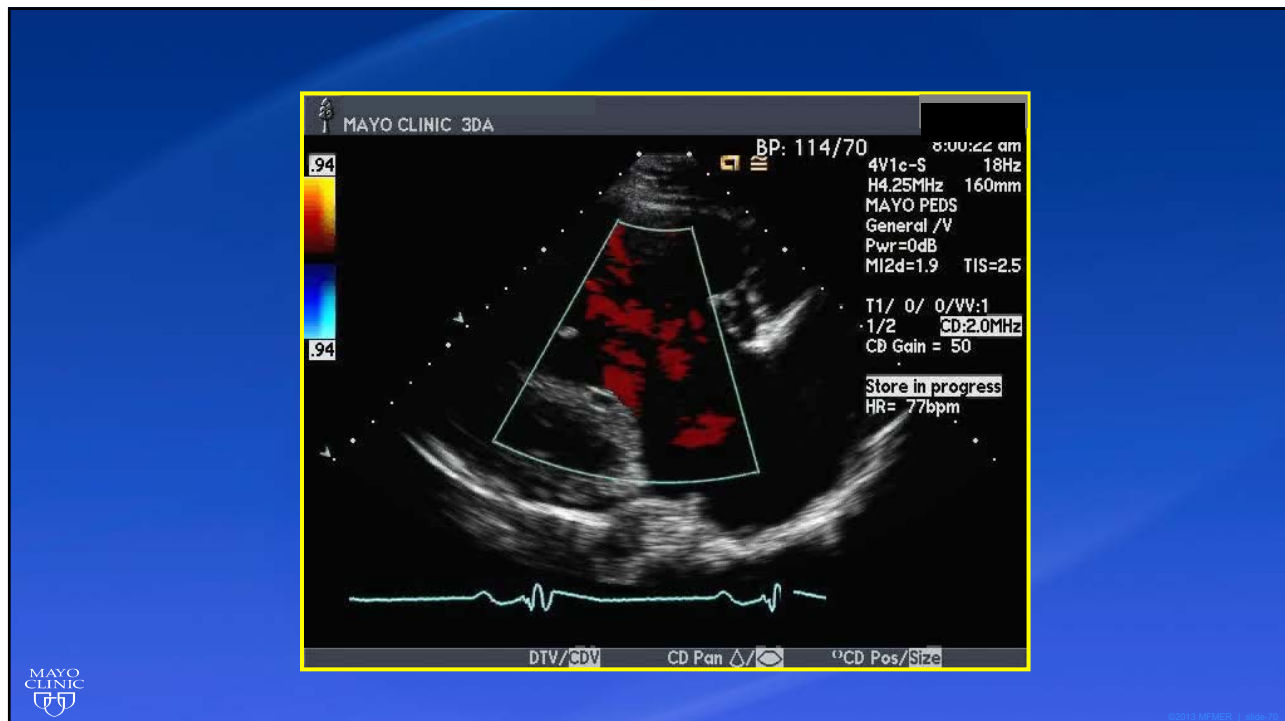
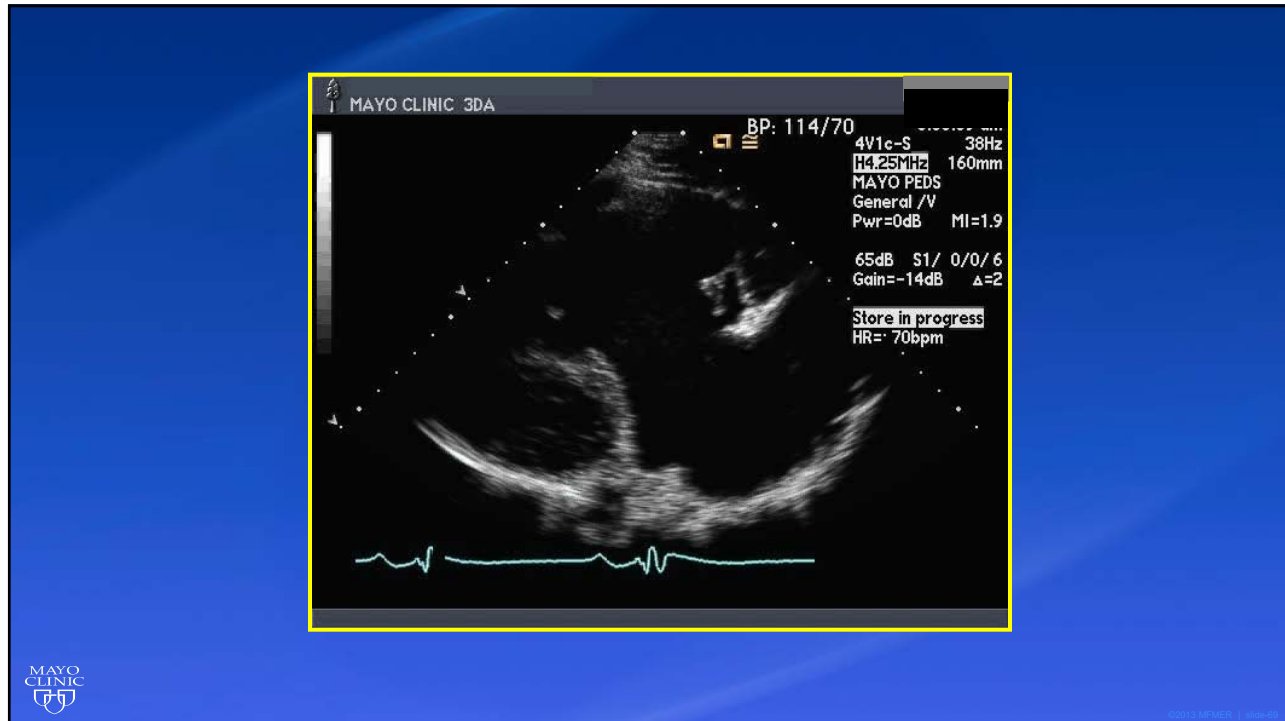
- Secundum ASD
- RV outflow tract obstruction
- LV non-compaction
- Accessory pathways

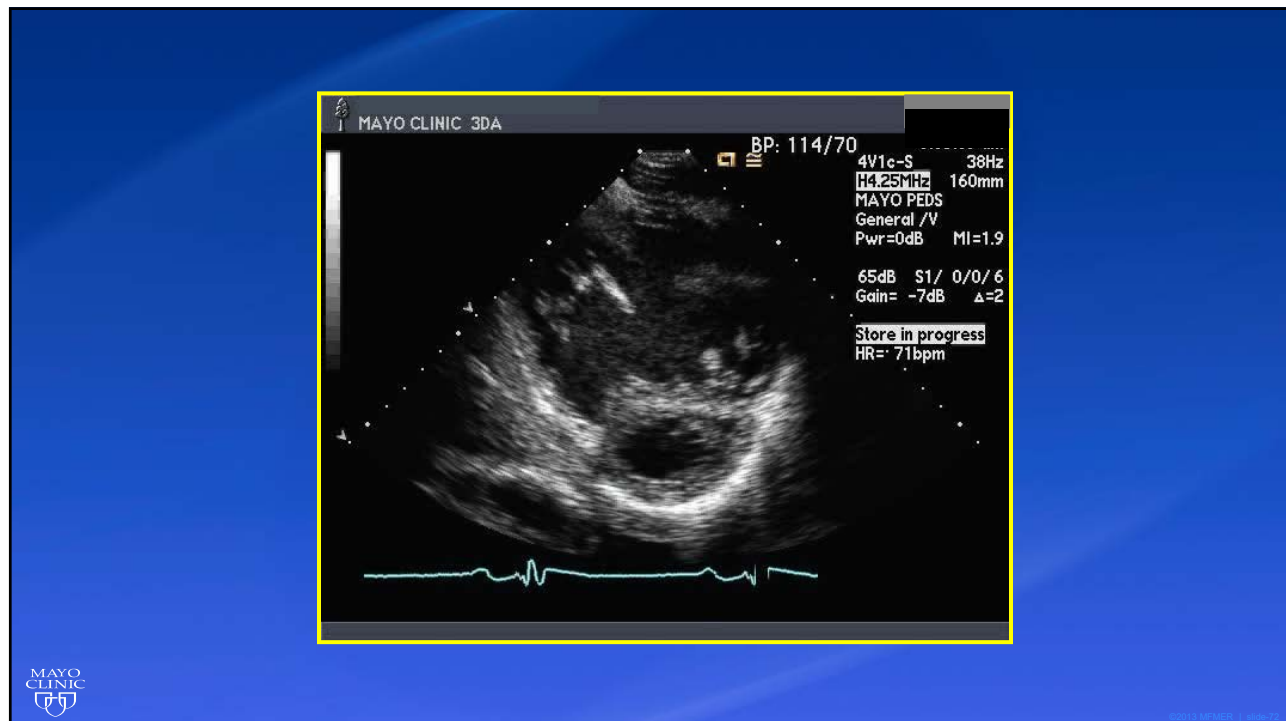
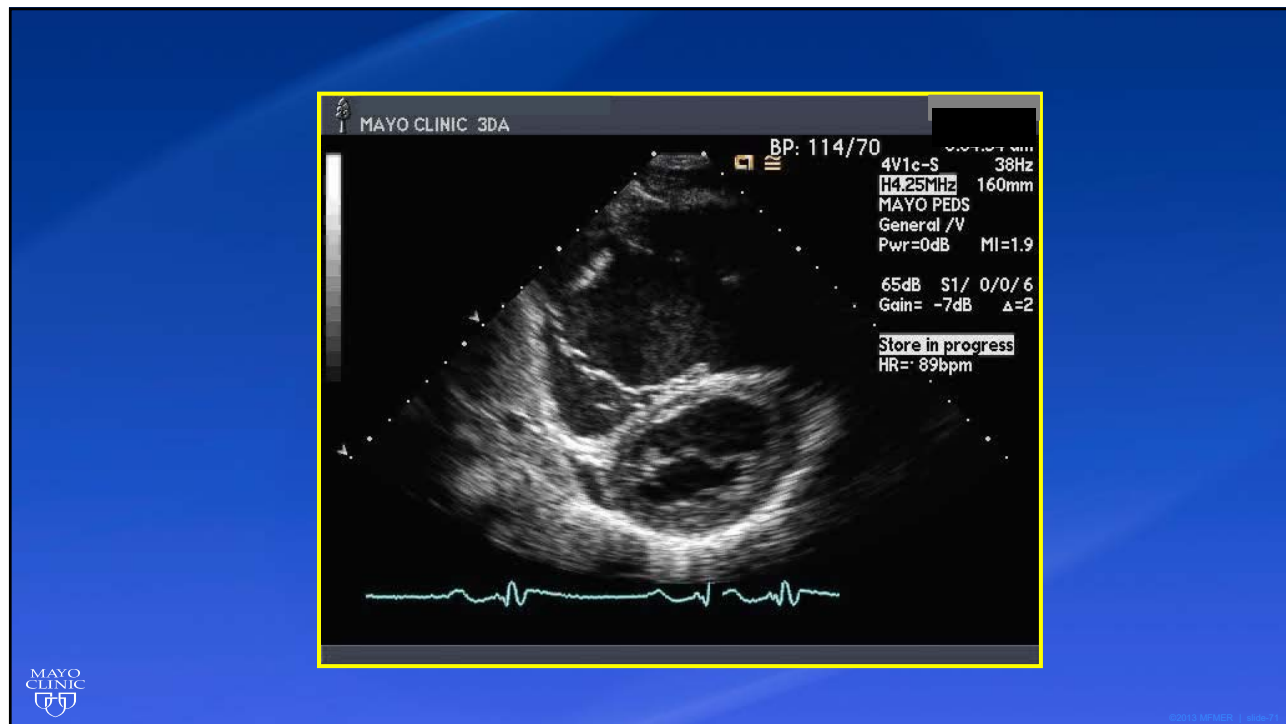


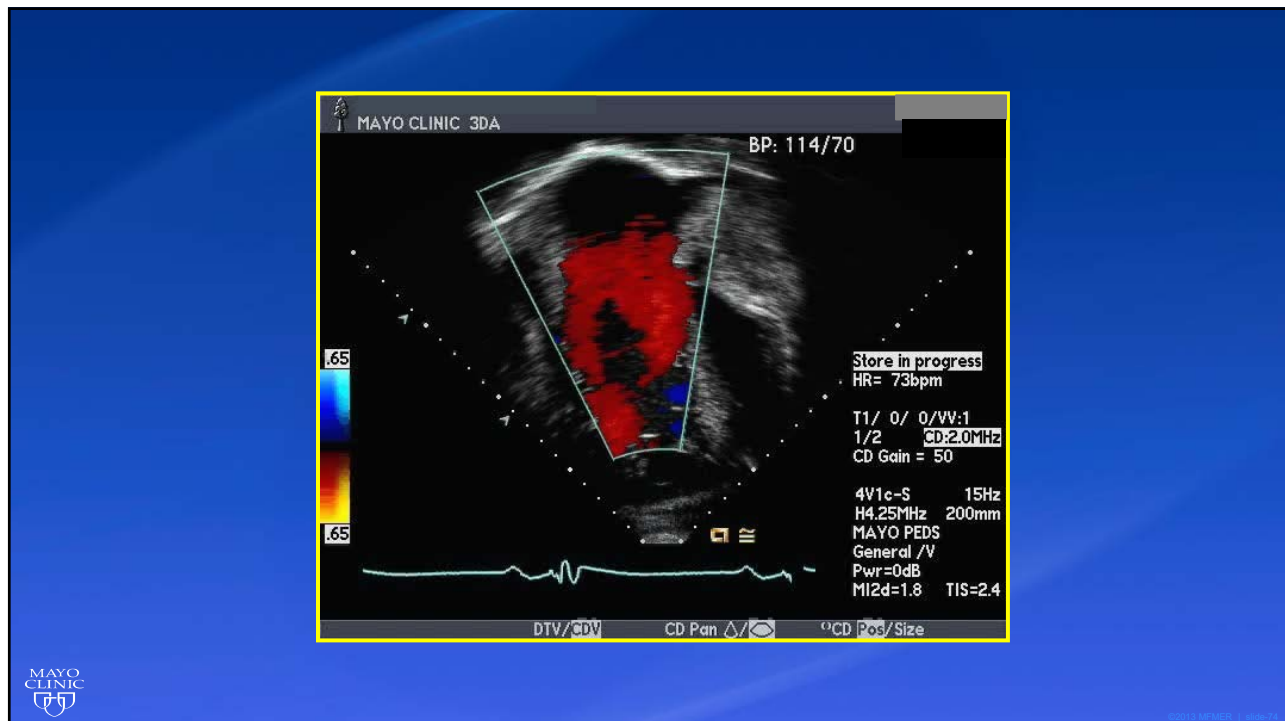
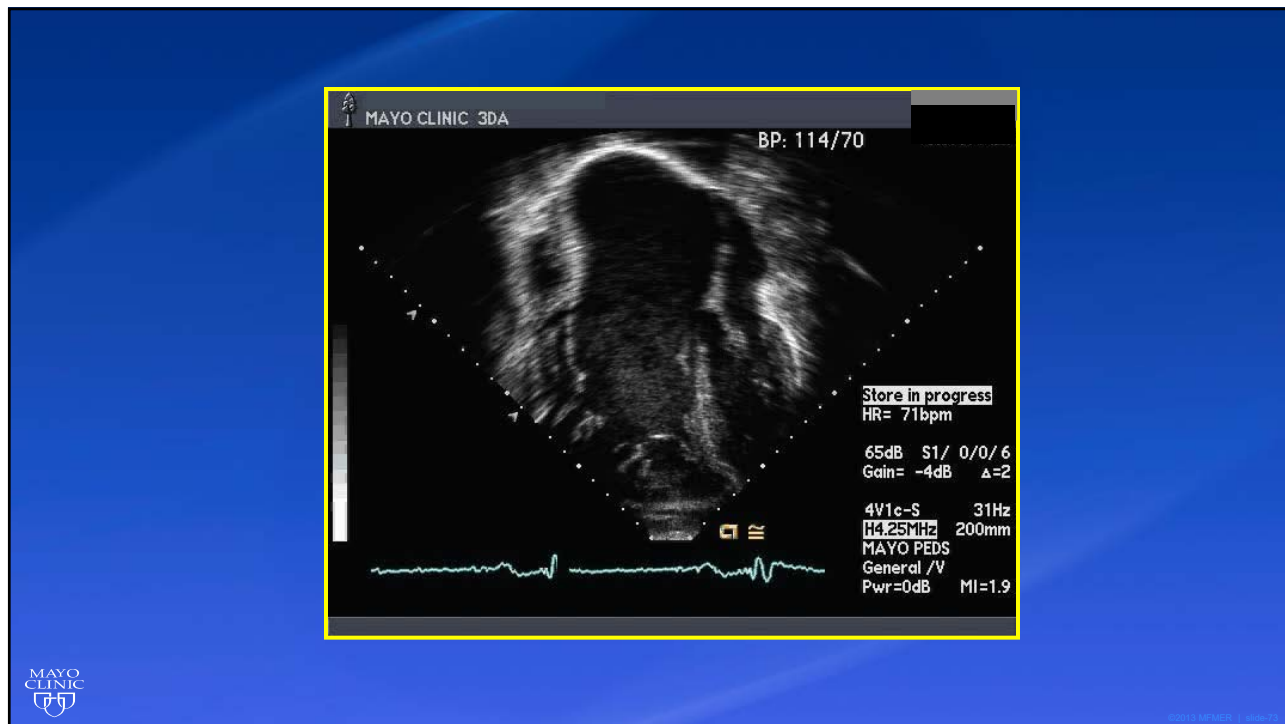
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Ebstein Anomaly

Indications for Operation

- symptoms, ↓ exercise tolerance, cyanosis
- progressive RV dilatation
- *before* significant RV dysfunction
- onset, progression of atrial arrhythmias
- ? earlier operation if TV repair is likely
- prior to LV dysfunction

Coarctation of the Aorta



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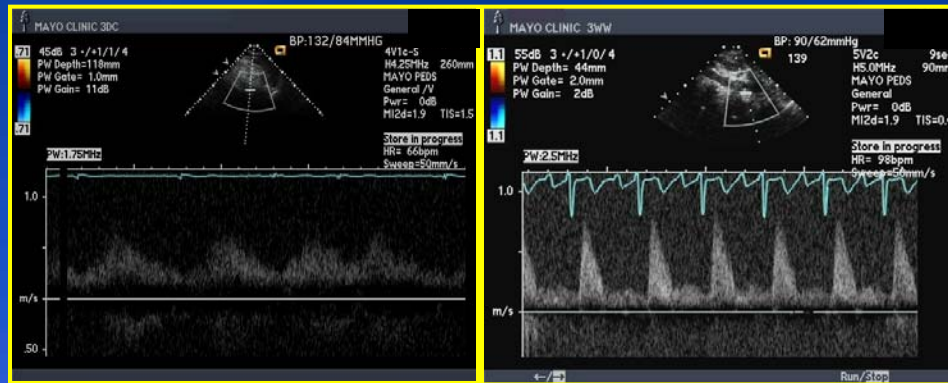
Imaging of Coarctation of the Aorta

- Abdominal aorta Doppler
- Suprasternal notch imaging
- Parasternal short axis - ?BAV
- Parasternal long axis – ascending aortic dimension



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Abdominal Aortic Doppler



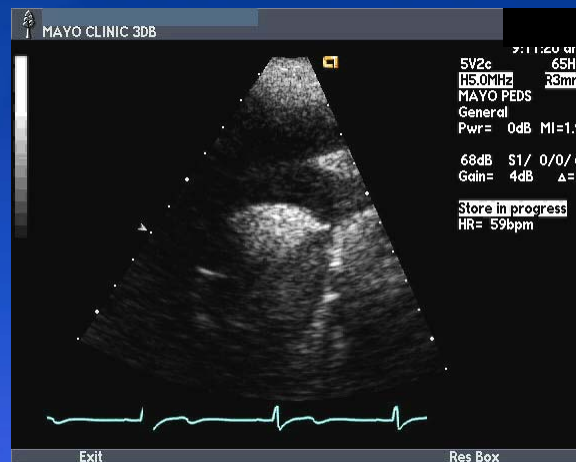
Significant Coarctation

Normal

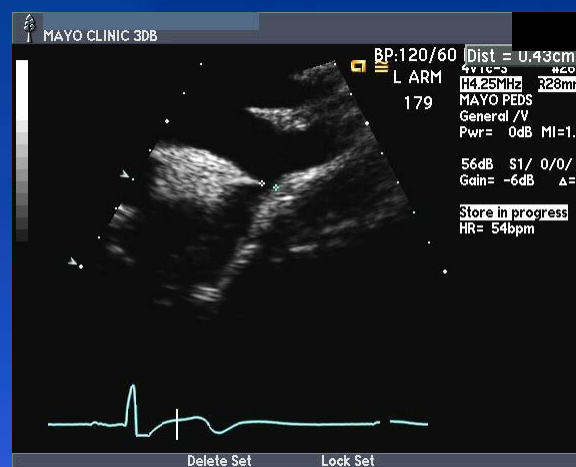
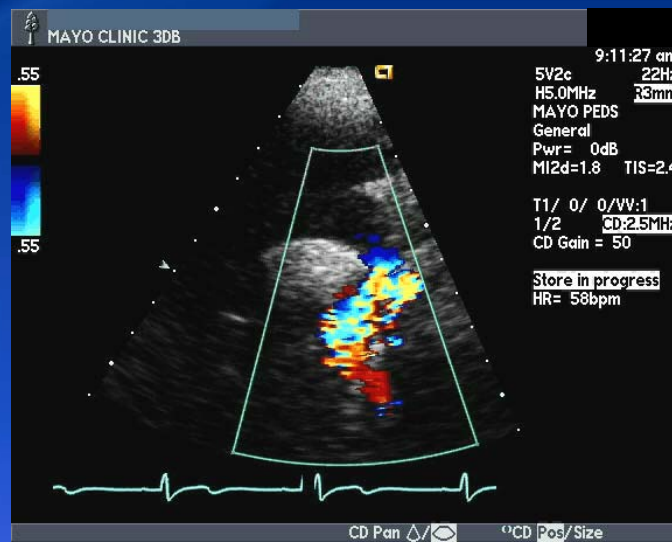


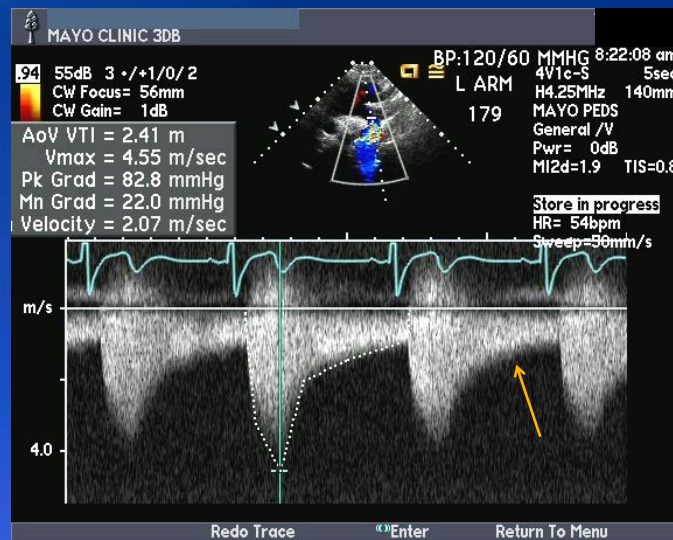
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Discrete Coarctation



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Coarctation Caveats

- Doppler gradient through the coarctation may be low 2° collaterals
- Abdominal Doppler pattern is critical
- Continuous flow in the thoracic aorta is helpful
- Don't forget association to BAV



Congenitally Corrected Transposition

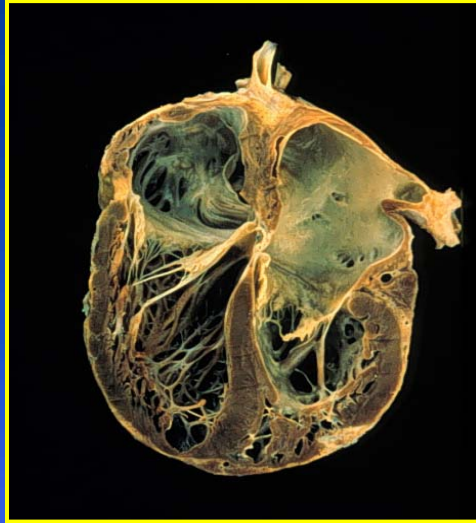


Image Courtesy of Dr. Bill Edwards



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Complete Transposition (D-TGA)

RA	LA
AO	PA
RV	LV

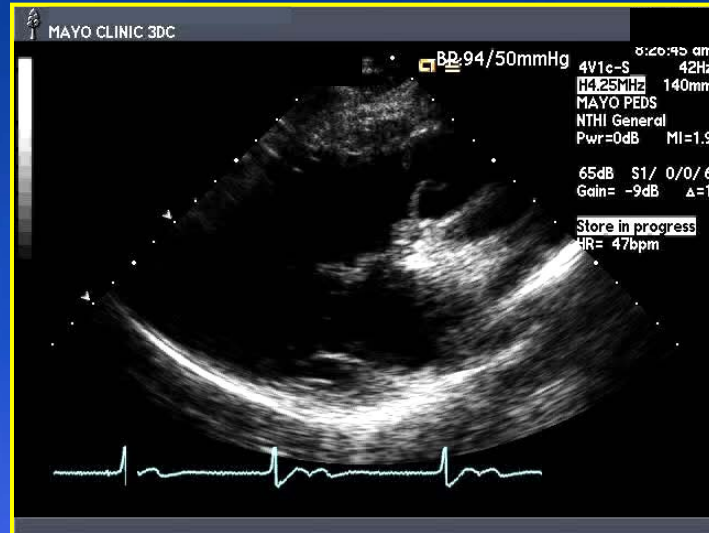
Congenitally Corrected Transposition (L-TGA)

RA	LA
PA	AO
LV	RV



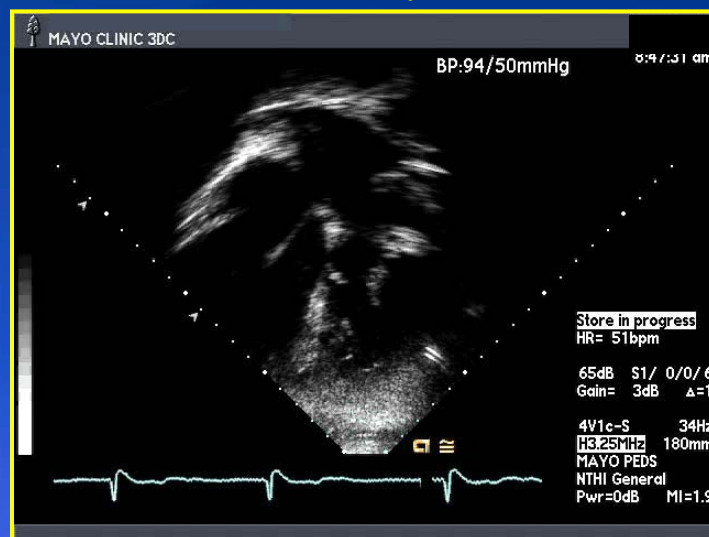
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Conus present in the "LVOT"



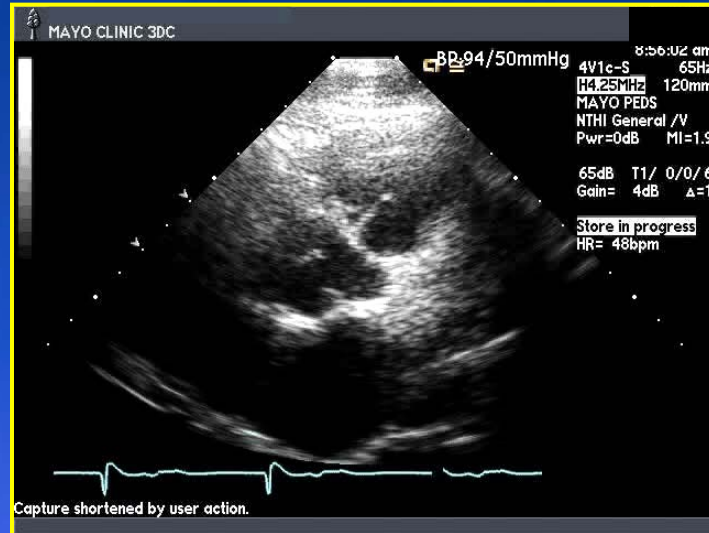
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Left A-V valve displaced apically



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Side-by-side semi-lunar valves



Lesions Associated with ccTGA

- Ventricular Septal Defect (70%)
- Subpulmonary ventricular outflow tract obstruction (40%)
- Tricuspid valve dysplasia/Ebstein malformation (90%)
- Situs Inversus
- Dextrocardia

Sequelae of L-TGA

- Systemic (RV) failure
- Systemic AV valve regurgitation
- Complete heart block
- SVT
- Sudden cardiac death



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Systemic AV Valve Regurgitation

- Surgical intervention is needed prior to significant decline in systolic function



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Conclusion

- There are congenital heart defects that present for the first time in adulthood, and they are not all “simple” lesions
- Right heart enlargement: ASD, PAPVR, TR, PR
- Left heart enlargement: VSD, PDA, AI, MR